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ART. I.—PROGRESSIVE FACIAL HEMIATROPHY.

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THE small number of recorded cases of the interesting affection known as progressive facial atrophy, with the consequent incomplete symtomatology of the disease, and the obscurity of its pathology render additional observations very desirable. The two following cases which have come under my observation, within the past six months, are apparently instances of this disorder, and as such they are therefore of interest. They present, moreover, certain peculiarities which, if they are not of service in clearing up the disputed questions in regard to this affection, are at least, somewhat suggestive as to some of the hypotheses that have been proposed in respect to its pathology.

Case I. The first of the two cases I have to relate, is that of Robert Berger, printer, aged twenty-two, of German parentage, five feet, five inches in height, weight about one hundred and twenty pounds, dark complexion, and apparently bilious temperament. No family history of nervous disease, and no previous illness since infancy, when he was said to have had typhoid fever. My attention was first called to him in June last, by his employer, who noticed that, in working a hand printing press in hot weather, he perspired only on the right side of his face, the left side remaining perfectly dry, no matter how warm the temperature or hard the labor, and that the line of demarcation between the wet and dry sides, was perfectly plain, and very conspicuous down the mesial line of the face. On my examination this was found to be the case, the perspiration being confined entirely to the right side of the forehead, the right cheek and side of nose and lips, on the chin, however, it encroached a little on the left, though not to any extent. Under the chin and in the throat, there was, he thought, a difference in the amount of the perspiration in favor of the right side, but not a very considerable one. On other portions of the body, there was no particular difference between the two sides in this respect.

At first sight, this looked like an ordinary case of facial hemidrosis, which is not so absolutely rare, but a closer examination showed quite a marked difference in the two sides of the face, and the patient voluntarily mentioned some peculiarities that could be observed on close inspection, and which he had himself observed. The left side was plainly less full than the other, the skin appeared tenser and more shining, the subcutaneous adipose tissue seemed to be largely lacking, the muscles did not seem to have lost any of their volume, but the hollows between them were more prominent,—noticeably so when he smiled. He stated, also, that a cut or scratch on the left side was longer in healing than one on the right. The beard as worn, was limited to a small moustache, which, though formerly black, was now reddish brown, with numerous gray hairs, which was likewise common in the hair of the scalp; and as he stated, had always been so. The moustache, so far as there was any difference in the two sides, was thinnest on the left, the hairs were continually falling out. The cheeks were kept closely shaven, but there appeared to be an equal growth on both sides. No paralysis, twitchings, or any disorder of molity whatever.

The left eye was a little less prominent than the other, the degree of enophthalmus though very slight, had already attracted the attention of the young man himself.

Careful measurements showed no appreciable difference between the two sides, none, indeed, could be expected at this stage when the affection had not yet advanced sufficiently to involve the muscles or bones.

The temperature was carefully tested in both ears, and a difference of six-tenths of a degree Fahrenheit, in favor of the right ear was found at the time of measurement, that of the left ear being 97° .6 and that of the right 98° .2. This was repeated on another occasion, and with nearly the same result.

The secretions, apart from that of the perspiration, already mentioned, were examined or inquired about with particularity. The tears were said to be secreted normally on both sides, the same was the case with the secretion of the mucous membranes of the nostrils, both in health and when suffering from a temporary catarrh. The salivary secretion was also undiminished on the left side; although the patient habitually chewed tobacco on that side, he did so more on account of habit, than, as he said, any convenience afforded by the lesser quantity of saliva.

Tactile sensibility of the two sides of the face was normal, or, if in any way at all altered, it was slightly increased on the affected side. A very careful use of the aesthesiometer developed nothing more. The special senses were all apparently normal, except that of taste, which was diminished or lost on the left posterior third of the tongue. It was tested with aromatics (peppermint, etc.) and with quinine. The former were very quickly appreciated on the left anterior surface of the tongue, and on the whole of the right side, but neither these nor the quinine were tasted at all, when placed on the posterior third of the left side until by dissolving and becoming diffused, they had affected the right half also, and then they were likewise tasted, to a slight extent, on the left side This was tried several times, and on different days, with always the same result. Taste did not seem altogether gone, but much impaired and retarded. The general sensibility of the tongue seemed in no way abnormal. The tongue was protruded in a straight line, and presented no atrophy, the palate was also normal. Electro-contractility of the muscles normal.

The first upper molar of the left side, was complained of as sometimes aching, and the gum was so wasted and the bone also, as to expose its root for a considerable distance. This was the only abnormal appearance observed within the mouth, and it had been noticed by the patient, only about four years, and the toothache, and occasional slight frontal headaches, to which he had been subject for many years, were the only painful affections co-existing with the nutritive trouble. The skin showed brownish freckle-like blotches, appearing, however, alike on both sides of the face.

The history of the case as far as could be learned was as follows: About two years ago, or perhaps a little more, the patient noticed that he perspired most on the right side of his face, and this gradually increased till at the present time the hemidrosis is more pronounced than ever before; it is in fact almost absolute, a small tract on the chin only being excepted. Lately, within the past year, he has noticed the slight inequality of the two sides, and the sunken eyeball. No injury can be stated, dating back to the time of the beginning of the symptoms, that can be reckoned in any way as a cause; one or two slight injuries had been received within the last year or two, but the patient was confident that they had left no serious effects, and had no connection with his facial trouble. Thus within fourteen or eighteen months, he had received a blow on the right side of the head with a club, which lacerated the ear cartilage, and left its scar, but produced neither at the time, or afterwards, any serious consequences. At another time within two years he received a burn on the left cheek, but no perceptible scar remained. A possible cause may perhaps be found in the fact that, before taking up his present occupation, he had been engaged in driving an express wagon, and was subjected to much exposure in cold weather, and several times had his ears, nose, and cheeks, more or less frost-bitten. He was not inclined, however, to consider these as having anything to do with the symptoms he presented. He gave no history of any change of color in the skin at the beginning, and when questioned particularly, was positive that there was none.

Pressure on the cervical ganglia was without effect, either in producing pain or otherwise. The pupils were symmetrical, and presented nothing abnormal in their appearance at different times.

CASE II. Mr. E., merchant, aged forty-two, married, nervous temperament, light complexion, medium stature, consulted us in July last, on account of a series of nervous symptoms from which he had suffered for a considerable time previous. His health had been generally good, though not robust, and there was no family history of disease; his ancestors had been healthy for four generations back. Habits unexceptional.

In 1864 he was thrown from a stage-coach, and fell on his head, not losing consciousness but receiving a scalp wound over the coronal suture, a little to the left of the median line, which bled freely. The next day, however, he lost sensation in the scalp, and had occasional blurred vision, and when he fell asleep he became delirous, and remained so for about a month. At the end of this time when he recovered consciousness, he had a dull pain in the top of his head, occasionally in the occiput, which he had experienced more or less till the present time. There was also a feeling of confusion of mind, aggravated by any mental or physical exertion, especially the former. These symptoms were so severe that for two years he was obliged to retire from active business, and they are liable to recur at the present time, in case of unusual mental worry or excitement, or loss of sleep.

But without going further into the general symptoms of the case, which indicated, I think, a considerable circulatory disturbance in the brain combined with neurasthenia, I will pass at once to those presented on the side of the face. At first glance the symmetry was apparent, the left side being plainly smaller than the right, the atrophy affecting not only the soft tissues but also the bones. The facial muscles were all diminished in volume, but functionally as active as ever, those of mastication, the masseter and temporal, seemed, on the other hand, to be unchanged. The adipose tissue was also wasted. The skin appeared of the same color on both sides, a little thinner perhaps on the left.

Careful measurements gave the following results:

	R.	L.
From outer canthus of eye to angle of lower jaw, - - - -	5 in.	3½ in.
From insertion of ala nasi to tragus,	4½ in.	3⅞ in.

This narrowing of the left side of the face, gave something of an appearance that the face was drawn to that side.

The beard was only thinly developed on the left as compared to the right side, and the patient stated that he only had to shave on the right cheek, the left producing no hair at all. As far as could be learned the secretions were all normal, except that he did not ever perspire on the face to any extent; as far as he knew there was no difference between the two sides. The temperature of the two sides was apparently the same. Nothing abnormal was noticed within the mouth, the teeth, palate, etc., were as usual, but when the tongue was protruded it seemed to deviate a little toward the right, perhaps on account also of the narrowing of the face to the left.

Several peculiarities of sensibility were presented. The tactile sense was certainly somewhat affected on the left side of the face, the two points of the aesthesiometer could not be distinguished on the cheek of that side, when separated an inch or more. Hearing was also affected; with his right ear he heard the not very loud ticking of a watch at two feet, with his left he only heard it from twelve to fifteen inches. Taste was decidedly weakened over the whole left half of the tongue, as tested by aromatics, a fact that the patient declared he had never before noticed. The eyesight was good, it was not tested systematically, but there was evidently no difference between the two eyes. There was no enophthalmus.

The patient's own testimony, and that of several of his acquaintances, who had known him from boyhood, is that this facial inequality had all appeared within the last ten or twelve years. A copy of an old photograph also, taken twelve or thirteen years ago, rather imperfect, and otherwise unsatisfactory, being a quarter face view, does not show this difference as now exhibited, or anything approaching it. In both this and the other case, the history so far as could be obtained, excluded all possibility of any congenital malformation or accidental deformity; they both appear to be plainly cases of the trophoneurotic trouble of the face to which I have referred them.

The two cases I have related present quite marked differences, and are alike mainly in the general symptoms of unilateral atrophy of the face. The first case was one of the incipient stage of the affection, before the atrophy had become marked, or had involved more than the adipose tissues; in the second the trouble was of ten or twelve years standing, and not only the adipose tissue, but the muscles and the bones themselves were atrophied: in one case we have no apparent cause for the disorder, the senses intact, excepting that of taste, and unilateral sweating of the head and face; in the other we find a traumatic origin, the tactile, auditory, and gustatory sense impaired, and no secretory phenomena whatever. Both present peculiarities that distinguish them from all the recorded cases as far as known to me. I will proceed to review the principal features of these cases in more of detail.

In the first of the two cases no positive cause could be adduced for the disorder, unless the slight frost-bites from which the patient had repeatedly suffered, are to be considered as such. They are sufficiently common in this climate in the winter months to be reckoned of very little consequence by out of door workers, and yet they may have had a part, at least, in this case in originating the morbid process. Still we cannot attribute any positive share to them. The case is, also, not altogether singular in having no direct perceptible cause, the same was true in an observation of Guttmann (*Archiv f. Psychiatrie* II. 1868.) The illness in infancy, seventeen or eighteen years or more previously, is rather too remote to be considered in this connection. In the second of our cases, on the other hand, the fall on the head in 1864, with the serious symptoms which succeeded it, appears to be an adequate causal moment, and as such has its precedents in the history of the disease, in cases reported by Romberg, Emminghaus, and others. In fact other traumatisms than cranial injuries, seem to occasionally cause the symptoms known as progressive facial atrophy, since Panas has reported a case (*Gaz. des Hopitaux*, 1869,) in which the wasting of one side of the face, followed a fracture of the lower jaw. In our case the injury being on the left side of the head, seemed to have all the more a direct connection with the symptoms, and in its nature and location, it was very similar to the one reported by Romberg (Klin. Wahrnehmungen, p. 91, 1851,) and one of Schuchardt given by Stilling, (Spinal Irritation, 1840, p. 325) in which there was a scar on the sight parietal bone, an inch long, and a quarter of an inch deep, caused by a fall in infancy. In our case we can conceive the intra-cerebral lesion of the atrophy, to be a direct, and not a reflex one, as in the case of more peripheral traumatisms, or lesions of disease, such as syphilis or diptheria. The very serious and uncomfortable cerebral symptoms experienced by the patient after the injury, and more or less ever since, are such as are not infrequently met with without any symptoms of trophic trouble in the face; and the atrophy in the present instance may be considered as an incidental phenomenon to the general cerebral disturbance.

A symptom which is mentioned by nearly all writers as being usually one of the first to appear, is a local discoloration of the skin, whitish or brownish spots preceding the wasting process, due to the disappearance of the subcutaneous adipose tissue. In neither of our cases was this appearance mentioned by the patient, or his friends, and careful questioning failed to recall to their recollection any such phenomenon The trouble began more insidiously than usual or such preliminary symptoms were overlooked. The latter, I think, was very possibly the fact in our cases; the patients were both males, one constantly employed on active business, the other a young working man, neither of them inclined to look after their symptoms, while most of the cases recorded of this disease have been in women, who are much more likely to observe these points. The absolute lack of any pain which could be referred to the parts involved, enabled the changes to progress without the consciousness of the patients, and this seems to be true in many of the other recorded cases of this affection.

The two cases were likewise different as regards the temperature of the two sides. In that of the young man Berger, a difference of six-tenths of a degree Fahrenheit, was met with on two separate occasions, while in the other patient, at the time of examination, no difference was discovered, notwithstanding the much more advanced condition of the atrophy. Both of these conditions, temperature diminished, and temperature unaltered, have been repeatedly observed in this affection, and there seems to be no constant rule in regard to this, except that perhaps the temperature is more frequently normal on both sides than otherwise.

The hemidrosis in the case of Robert Berger, is also of interest, since it has been observed but seldom in any degree in this affection. In our case it was almost complete over the head and face, and existed to a certain extent on the neck and throat. In the other patient, the absence of perspiration on the head and face throughout, is a noticeable feature, but its connection with the atrophy is not clear. The significance of the hemidrosis is also obscure, but its coincidence on the same side as the nutritive trouble is more suggestive. Nicati (Sur Paralysie du Nerf Sympathetique cervical. Lausanne, 1873) considers a combination of symptoms, several of which were present in this case; lowered temperature, stoppage of perspiration, and wasting of the side of the face, as indicating a paralysis of the sympathetic nerve, in what he calls its second or more advanced stage. But here other phenomena which we would look for, were this the fact, are lacking, such for example, as pupillary contraction, drooping of the eyelids, etc., and the symptoms of a preliminary stage are altogether wanting in the history of the case. Brunner (*St. Petersburger Med. Ztschr.* II. 1871,) gives an account of a case in which the perspiration was decreased, as were also the tears and mucous secretions, the temperature diminished on the affected side, and pain was experienced on pressure over the upper cervical ganglion. There was also exophthalmus, and the pupil was dilated on the side of the atrophy. In this patient, according to Brunner, there was a permanent irritation of the cervical sympathetic by a tumor. In our patient, on the contrary, there were none of these symptoms of irritation of the sympathetic.

While we cannot exclude the possibility of the participation of the sympathetic in this case, the evidence is not quite sufficient to positively assert it. The perspiration may be controlled by special secretory fibres, as is held by many physiologists of the present day, and these may be involved in the present case, or its stoppage may be due to a mechanical interference with the sudoriporous glands or their involvement in the course of the nutritive trouble. The extent, however, of the surface over which the sweat is stopped or decreased, is much greater than that implicated in any degree in the atrophy, and this latter supposition seems therefore, hardly probable. It appears to me that we must prefer the other hypothesis, that the centres, peripheral or central, or in the sympathetic, of the secretory nerves, are involved, in the production of this phenomenon.

The sensory symptoms in both of our cases are worthy of attention, and, indeed, are among the most interesting of all that are presented. They are so in the first place because disturbances of sensibility have been but rarely observed in this affection, and, secondly, because some of the phenomena noticed in these two cases are entirely new in the history of the disease. A diminution of the general tactile sensibility of the skin of the affected side has, it is true, been observed by Tanturri (*Il Morgagni*, 1872, quoted by Eulenberg in Ziemssen's Hdbch, X. 2, and by Vulpian (L'Appareil Vaso-Moteur. II. p. 430) and perhaps by others. In the case of Mr. E. it was quite plainly diminished on the atrophied left side. The sense of hearing, which was likewise affected in him, has not been observed to be noticeably impaired in other cases of the affection, at least not in any way as connected with it. In our case the impairment of hearing was slight, and it may *possibly* have been due to causes totally unconnected with the facial trouble. An examination of the ear, however, revealed no such cause, the patient was not aware of any, and I am, therefore, inclined to think that it was one of the nervous symptoms following the injury which produced his facial trouble, and the cerebral disorders accompanying it. He was moreover, especially liable to vertigo, induced by the slightest causes, such as suddenly turning around, etc., and this too, may possibly indicate trouble in the internal ear. But allowing all this, there may be no necessary connection between the atrophy and the deafness, they may be only coincidences though due to the same general cause.

The most interesting phenomena of sensibility, in the two cases I have related, are those presented by the sense of taste. In both it was impaired, in Mr. E. the whole left half of the tongue was involved, in the other only the left posterior third. In the first of these, the functions of both of the glosso-pharyngeal and the chorda tympani were affected, indicating the implication of both these nerves. The significance of this observation is that either the glosso-pharyngeal and fifth nerves, are both involved, or that the taste fibres for the base as well as for the tip of the tongue are derived from the last named nerve. There seems, however, to be no necessity for adopting this latter view in this case, the general cerebral disturbance caused by the traumatism, the implication of the hearing, etc., seem rather to indicate at least, an equal probability of an intracranial lesion affecting to some degree the nucleus of the glosso-pharyngeal. It favors, however, the theory of Schiff, as to the course of the taste fibres of the chorda tympani, through the trigeminus to the brain, since there are no evidences of any involvement of the facial nerve.

In the case of Berger, the condition is still more interesting. There was no traumatism, causing general cerebral disturbances, and rendering possible the assumption of lesions of the nuclei of various cranial nerves; the facial atrophy was still only slight; there were no other sensory symptoms, or pain, except the aching of the molar mentioned in the account, but the sense of taste was very seriously impaired over the left posterior third of the tongue. While quinine, placed on the right side, was tasted almost at once, when placed on the left the young man was almost unaware of its presence, after a minute had elapsed, and until its diffusion by movement, or by the saliva, no bitter taste whatever was felt. At the same time, sweet and aromatic substances were appreciated alike on both anterior halves of the tongue. The experiments were repeated several times on different occasions, with the same result. I have no good reason to suspect any deception in this case; the young man's statements were positive, and I believe him truthful, and he certainly gave no signs of tasting the drug.

It is hard to account for this condition of affairs by any theory that has so far been advanced, either of the disease, or of the physiology of taste. The atrophy seemed to involve the region supplied by the second division of the fifth nerve as much as, or more than the other divisions; and on this is situated the spheno-palatine ganglion which gives off the vidian nerve, the upper division of which, the superior superficial petrosal nerve, is supposed by Schiff to contain the taste fibres, which return from the lingual nerve through the chorda tympani. The usual theory of this disease, which I shall review later, is that it depends upon an affection of the trophic fibres contained in the fifth nerve, or one or more of its divisions. Accepting this theory, Baerwinkel (*Deutsch. Archiv. f. Klin. Med.* XVII., I., 1875,) noticing a case of unilateral atrophy of the face, affecting only the second division of the fifth, and, as he held, indicating trouble with the spheno-palatine ganglion, its trophic centre, mentions the fact that there was no impairment of taste in the anterior portions of the tongue, as rather against the hypothesis of Schiff, as to the course of the taste fibres. At the same time, he gives two other observations of lesions of the trifacial and the seventh nerve that favor it. In our patient, I should have expected *a priori* to have found the taste affected in the anterior portion of the tongue, if anywhere, but the reverse was the fact. It is difficult to suppose an accidental lesion of the glosso-pharyngeal co-existing with the one of the trigeminus that produces the atrophy, when there are no more signs of nervous disorder than there were in this case. On the other hand, the taste fibres of the glosso-pharyngeal are not generally supposed to have such connections with the fifth nerve as to be involved in its disease, while those of the chorda tympani, of the connections of which with the trigeminus there is much more physiological evidence, escape. The observation is a contradictory one, and I cannot, at present, explain it.

It is worth noticing in this connection, that neither of the two patients were aware that their sense of taste was impaired previous to my examination, the opposite half having, I suppose, supplied all the gustatory sensation of which they experienced the need. And this makes it seem possible that similar conditions, unreported by the patients, may have existed in other cases of this disease, though none have been recorded. It is not stated in all the observations, that actual experiments on this sense were performed; and if merely the patient's testimony was accepted, there was plainly an opportunity for error. We are not conscious, unless we give the matter especial attention, that we taste sapid substances on one side or the other of the tongue. The sensation is generalized over all parts, and it required, as is well known, special experiments to determine what portions of the gustatory surface appreciated the most opposite sensations, or what were really the parts of the oral mucous membrane that participated in these sensations; and, finally, we have to consider the subjective confusion between the gustatory and olfactory sensations derived from certain substances. All these facts show the necessity for careful examination and actual experimental tests, in deciding as to the condition of the sense of taste.

The general tactile sensibility of the tongue seemed all right in the case of Berger; no noticeable difference between the two sides was detected. In the other patient it was not so thoroughly tested, but seemed normal. There was no opportunity for a subsequent examination in his case, his residence being at a distance.

One other point in the case of young Berger appears to deserve especial mention; the exposure of the root of the first upper molar, on the left side by wasting of the gum, and alveolar process, in front of it. The root was exposed nearly its whole length, a third of an inch or more, but appeared otherwise sound. I find by inquiry among dentists that such a condition is tolerably rare in young persons, though now and then met with in older ones. When it does occur in young individuals, it is generally attributed to a congenital deficiency of the alveolar process in that special region, which sooner or later reveals itself in this manner, or to continuous pressure from some cause or other. In this case there is an appearance of an enlargement of the root; the tooth is painful on pressure, and it is said to have been noticed in this condition only about four years. There may be no real connection between this dental peculiarity and the incipient nutritive disorder of the face, but the coincidence of the two is certainly worthy of note. The young man is a habitual chewer of tobacco, and this may be the cause of the phenomena.

Having thus reviewed the most prominent features of these two cases, I will pass to notice their bearing of the theories that have been offered of the pathology of the disorder. Nearly all authorities admit the neurotic character of the affection; even Friedreich in the preface to his great work, on progressive muscular atrophy, in which he maintains that this is a disease of the muscular, and not of the nervous system, recognizes these partial atrophies of the face, even when connected with the atrophy of other parts, as a limb, etc., as neuroses. Only one author, I believe, has recently proposed an opposite theory; Lande in 1870, who held it to be an affection primarily of the connective tissue, acting secondarily and mechanically on the nutrition of the other tissues by interfering with the circulation. Very few, however, have adopted this theory; the nervous phenomena which so often accompany the disease, and are undoubtedly directly connected with it, and its obvious neurotic cause in many cases, such as those in which it follows a cerebral injury, as in one of the cases, I have related, are sufficient, I think, to invalidate it. As Eulenberg (Ziemssen's Hdbch. XII. 2, 69) says, it has only the standing of a hypothesis unsupported by anatomical proofs, and strongly opposed by clinical evidence.

Two quite different theories of the neurotic character of the disorder have been advanced; one that it depends upon an affection of the vaso-motor system, affecting in this manner the nutrition of the parts; and the other stated in a general way, that it is due to an alteration of the trophic influences conveyed mainly through fibres contained in the trunks of other, chiefly sensory, nerves, and in the case of this affection, especially in the division of the fifth nerve. The vaso-motor theory is maintained, I believe, by but few authorities at the present time. In the first place, while we may not be able to always absolutely exclude the sympathetic system from some participation in the production of the symptoms presented in this affection, as a whole they are very different from those we observe in disorders, experimental and clinical, of this system. In fact it is mainly the phenomena of neurotic atrophies, that have led physiologists to assume the existence of special nerve fibres influencing nutrition—the trophic nerves of Samuel—which if not now universally, are yet very generally accepted as physiological facts, inferentially proven, though not anatomically demonstrated.

I have said all that is perhaps necessary to say in regard to sympathetic involvement in my cases, when speaking of the unilateral sweating of young Berger. The influence of the fifth nerve in these cases still requires to be discussed, to see how far it supports the prevalent theory of the disease. In the first case, that of Berger, the atrophy, as far as it was perceptible, seemed to affect all parts of the left side of the face, the eye was slightly sunken, the cheeks wasted, and it could be detected, I thought, also on the chin. Two points, however, do not support the trigeminus theory, though they are not conclusive in any direction, they are the unilateral sweating which certainly extended over a much greater surface than is supplied by the fifth nerve, and the affection of taste, which I have noticed, as being difficult to account for. I cannot add any remarks here on these points of the case. In the second patient, Mr. E., there was, on the other hand, no enophthalmus, or other indication of any involvement of the first division of the trigeminus; the trouble as far as this nerve is concerned, was apparently confined to the superior and inferior maxillary divisions. But it is not uniform over the region supplied by these branches, the tongue was symmetrical, and while the facial muscles generally were wasted, the masticatory muscles, except in so far as the diminution in size of the bones necessitated it, were apparently undiminished in volume. The paralysis of the chorda tympani, as regards the sense of taste, though never observed before, is not at all at variance with the physiological ideas of the function and origin of this nerve; that of the glosso-pharyngeal and the impairment of hearing on the contrary, is only to be accounted for by the general disorder caused by the injury to which the atrophy also is attributed. And these general symptoms of cerebral disturbance with their very obvious cause, affect the value of this case in aiding in the study of the pathology of this particular affection. It, by its limitation to the two lower divisions of the trifacial, nevertheless, so far, favors the opinion that this nerve is specially involved. The other case is, perhaps, still less decisive in favor of any one theory of the pathology of the disorders; one point may, however, be mentioned here, as having a possible bearing on this as well as its causation in this particular case. The dental peculiarity described, which first appeared some four years ago, and which is not common in young persons, may have had an influence in a reflex way in producing the symptoms. This is suggested only as possible, I would hesitate to pronounce any positive position. But, since it is well known that very insignificant appearing dental troubles may produce very serious effects in the form of neuralgias of the fifth nerve, often accompanied with obvious physical symptoms, it does not seem altogether impossible that similar causes might occasionally give rise to other manifestations of derangement, such as trophic phenomena in the region of this nerve. I offer this merely as a suggestion to be taken for what it may seem worth. There are, it is well known, trophic troubles that accompany, and are connected with neuralgias, and they have been studied by Brown-Sequard, Nothnagel (*Archiv. f. Psychiatrie* II. 1869.) and Vulpian (Lecons sur l'Appareil Vaso-Moteur, II. p. 435.) The latter author, very properly, I think, rejects the vaso-motor theory of these atrophies proposed by Nothnagel, and supposing the neuralgias themselves to be central, the central irritation which produces them may extend, and produce a kind of inhibitory action over nutrition of parts, by causing a kind of suspensory action on the spinal centres that control this process. Hence he objects to the term reflex, which has been applied to these atrophies, since they depend only on a diminution of the activity of the spinal trophic centres, and not on any centrifugal influence sent out over the muscles. This may be the proper way to explain trophic nervous action, and it certainly favors the view that it is confined to the sensory or centripetal nerves, but it nevertheless appears to me to be too fine a distinction. It is not easy to understand how the trophic centres in the chord can influence the nutrition of distant parts, without inferring the existence of centrifugal conduction of some kind or other, which might, together with the peripheral origin of the trouble in many cases, bring the whole properly under the head of the reflexes. Eulenberg, (l. c.) after mentioning the observation of Axmann, in whose patient the atrophy was preceded by spasmodic contractions of the muscles of mastication and hyperæsthesia of the affected side, and remarking on the probable extension of the irritation to the trophic fibres, says: "In this connection some weight must be laid on the fact that cases are not at all rare in the regions supplied by other branches of the trigeminus (especially its first division) in which sensory irritative phenomena, super-orbital neuralgias, for example, are conjoined with nutritive disturbances of the hair and skin over the track of the affected nerve. The transition between such cases as have been observed by Romberg, Anstie, myself, and others, and certain cases of circumscribed atrophy is very gradual, and with this conjunction of neuralgic and trophic disorders, a case may be considered either as one of super-orbital neuralgia, with trophic changes, or as one of limited local atrophy with neuralgic symptoms, according to the preponderance of this or that symptom, or the subjective ideas of distinction of the author." The above quotation expresses as well as I can state, and with the weight of the authority of Prof. Eulenberg, the close connection between these two manifestations of nerve disorder. I would only add to it, that it seems not altogether impossible that nerve disturbances not necessarily accompanied by neuralgic symptoms, such as sometimes occur in carious teeth where the nerve may be exposed and involved in morbid growths without pain, referred to this source, may also induce this condition.

Although the cases I have related, supposing them to be genuine instances of progressive facial hemiatrophy, which seems to me almost beyond doubt, do not throw much light on the pathology of the disorder, still, they do not strikingly contradict the opinion that the affection is one involving chiefly the trifacial nerve—the one which, I think, has the greatest support from clinical evidence, and the most theoretical probability. They certainly do not especially favor any other that has been advanced.

Allowing, then, the principal lesion to be in the trigeminus, it is worth while to theorize, and see if it cannot be brought within still narrower limits. There are many facts that seem to show that trophic changes often depend on alterations of the ganglia of the posterior roots of spinal nerves, and of the gasserian ganglion of the fifth, which, considering it to be a mixed nerve, is the ganglion of its sensory root. The facts of this kind are all, or nearly all, pathological, and the trophic troubles observed have been of a more active kind than those we have been discussing, still, the analogy has been used for the building of the hypothesis, that in the case of the disorder before us, the gasserian ganglion is affected. Baerwinkel has gone still farther, and, as has been already noticed, has considered that other ganglia, the spheno-palatine, for example, on the second, are trophic centres for the different divisions ot the fifth nerve. The difference between such changes as zoster, etc., which have been observed with ganglionic lesions, and the dystrophic, or atrophic alterations in the affections under consideration, are objections to this view, but are nevertheless not absolutely conclusive. As the disease has never been fatal, and but few cases have been observed, the opportunity for a *post mortem* examination has never been as yet afforded. I may perhaps be allowed to make a suggestion that occurs to me, that since the dystrophic changes in senile atrophy, though more general, often bear a considerable resemblance to those in this affection; a careful microscopic examination of the ganglion cells in the gasserian and other cranial ganglion should be made, and that if done it might perhaps throw some light on the dark questions of pathology we have met with. Dr. H. C. Major, (*West Riding Lun.*

Hosp. Reports, IV., 1874,) has investigated the condition of the cortical nerve cells in senile atrophy, and made some interesting observations; but no one, so far as I know, has attempted this for those of the basal ganglia, the medullary nuclei of the cranial nerves, or their ganglia. The field seems worthy of being worked.

That the morbid process in this disorder, as regards the tissues of the face, is purely atrophic, is shown by the retention of the voluntary and electric contractility of the muscles, and especially by the fact noticed lately by Hammond, (Diseases of the Nervous System, 1876, p. 550) that there is no degeneration, but simply a reduction of volume in all directions of the affected fibres. This lesion, if it can be called such, is entirely different from any that have as yet been observed in connection with lesions of the gasserian, or other ganglia.

Dr. Hammond does not accept the view that this disorder of nutrition depends upon an alteration in the fifth nerve, but attributes it to "an affection of the trophic cells of the bulb which are the nuclei of the facial, the hypoglossal, and the spinal accessory nerves." The grounds on which he bases this opinion are to be found in the resemblance between this affection and progressive muscular atrophy affecting the face, which disease the majority of authorities attribute to alterations of motor nerve cells in the anterior tract of gray matter. There are several difficulties, however, in the way of accepting this view, as to the seat of the disease. The alterations of the tissues are not altogether the same as, or similar to those of progressive muscular atrophy; they are more general, involving a greater variety of tissues, while, at the same time, they are less extensive, and do not ever go on to actual degeneration of muscular fibre, etc., as far as has been observed. The difference is one of quantity altogether, and not of quality; the morbid process appears to be slower, and of a more passive kind than that in progressive muscular atrophy. Then we have the limitation of the disease to special regions, innervated by particular divisions of the fifth nerve, and the gradation alluded to by Eulenberg, between neuralgic atrophy and atrophic neuralgias, both of which points have been previously mentioned in this article. Lastly, some cases, and among them the ones I have described, show the implication of still other nerves than the ones mentioned by Dr. Hammond; and in their case a functional involvement, which is more than is shown in any case by the facial, the hypoglossal, or the spinal accessory.

It is not at all impossible, it must be admitted, with the evidence we have at present, that there may be multiple lesions of the nuclei of various cranial nerves. There are, moreover, on record, cases of hemiatrophy of the face, co-existing with atrophy of other parts—an arm, or leg, for example. Such cases are, also, distinguished from progressive muscular atrophy by as high an authority as Friedrich, who, as already stated, admits their neurotic character. This being the case, it would seem necessary to admit either multiple lesions of trophic centres connected with both cranial and spinal nerves, or a disturbance at some higher common trophic centre controlling the nutritive functions of both. There is perhaps some room for doubt as to the correct reference of these cases to this disease. I have mentioned them only because, if correctly referred, they show the extensive range of nerve nuclei we must consider involved, or admit a still higher seat.

It may appear almost useless to attempt, with our present knowledge, to give any formal statement of the probable pathology of this affection. The actual lesions on which it depends, can, of course, be only certainly known after careful autopsies of many cases. The following, however, are offered as presenting briefly what seem to me the principal facts and probabilities in regard to this affection.

1. Progressive facial hemiatrophy is a neurosis. This is indicated by its limitations to one side of the face, and to the regions supplied by special nerves; by the implication, in some cases, of the sensibility, and even special senses; by the merely dystrophic character of the changes it produces in the tissues without any indications of disease, or alteration of the essential structure; by the close analogy in some respects with the neuralgias, attended with local atrophies, etc.

2. As regards the nature of the nervous trouble, it appears, from an analysis of the symptoms in a majority of the cases reported, that the essential lesion is not in the vaso-motor system, but rather in the trophic functions of other nerves. Vaso-motor symptoms, properly speaking, are lacking in a majority of cases.

3. From the evidence afforded by a large proportion of the reported cases, it seems justifiable to suppose that the trophic functions of the fifth nerve are especially implicated. The facts that render this probable, are the frequent limitation of the atrophy to the regions supplied by this nerve, or its separate divisions, the analogy with the neuralgic atrophies, and the occasional sensory symptoms. I may add, also, that the lack of symptoms that would lead to the inference of implication of the facial nerve, is in favor of this view, at least, as assisting in a kind of diagnosis by exclusion. The hypothesis that the atrophy depends upon alterations in the ganglia on the fifth nerve, cannot be said to be very well supported by the facts in our possession, as to the results of disease of these ganglia, but it may be the correct one.

4. In some cases there are evidences of positive lesions of other cranial nerves than the fifth; paralysis, or irritation of the sympathetic, (Seeligmueller, Brunner) paralysis of the abducens (v. Graefe), of the auditory and glosso-pharyngeal (cases reported in this paper). In some of these observations, where the disorder was apparently due to traumatic causes, syphilis, etc., the facial atrophy is perhaps to be considered as only one of the manifestations of the more general intra-cranial disease, of which these other phenomena were also symptoms. Still, there are some facts that indicate an alteration of nerve nuclei in the medulla, in many of the cases, the direct cause of which is not obvious.

5. The symptoms of the disorder indicate a chronic trophic asthenia or paralysis, rather than any irritative action. This is proven by the slow progress of the disease, its usual unirritative character, and by such symptoms as that testified to by one of the patients in the cases here presented; *viz.*, that slight injuries, wounds, scratches, etc., were longer in healing on the affected side than on the other.

6. That the disorder, however, is not merely one of arrest of development, is shown by the actual wasting of the tissues, the more solid ones, such as the bones, among them in many cases. In this, it has a resemblance to the atrophy of old age with which it seems to have other analogies, suggestive in regard to its pathology. The cases of congenital facial atrophy that are not infrequently met with, have not generally the progressive character of this affection, and are more properly to be reckoned as due to arrest of development. Still their relations with this disorder may be closer than we think.

7. There are various peculiarities of this disease which, in the present state of our knowledge, cannot be accounted for in any way. Still they serve to support the neurotic and local theory of the affection. Such is its unilateral character, the greater frequency of its occurrence in females than in males, etc.

8. Therapeutic measures have so far, failed to produce lasting benefit, for the most part. Two or three cases seem to have been benefited by faradization (two of Baerensprung) and the prognosis, as regards recovery or arrest of the disease, is not altogether hopeless. If, as has been suggested as possible, in one of the cases here related, it ever depends upon a reflex cause, or when it seems due to specific disease, like syphilis, the cutting off of the original irritation or proper specific treatment would seem to be the most promising remedial measures.

9. The disorder does not seem to involve life. It is possible, however, that in some cases it might extend to organs that are essential to life, especially if the nuclei of various cranial nerves are involved in the morbid process.